

THE TREATMENT OF PULSATING EXOPHTHALMOS.¹

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THE disease known as pulsating exophthalmos is the result, in the vast majority of cases, of an arteriovenous aneurism of the internal carotid artery and cavernous sinus, in the other cases, which are extremely few, it is due to an aneurism of the ophthalmic artery in the cranium. In about 71 per cent. of the cases it follows traumatism, in the remaining 29 per cent. its spontaneous origin is due to an existing chronic endarteritis of the internal carotid. Immediately following rupture of the artery, blood escapes into the sinus, and the pressure in the sinus is raised to a degree depending on the size of the rupture. This increase of pressure causes an interference with the venous return through the superior ophthalmic vein, and a state of passive congestion in the territory of the vein follows. The symptoms indicative of passive congestion may develop rapidly or slowly, according as the opening between the artery and sinus is large or small, and as a rule they appear early, but it is some days or weeks before they are fully developed. The characteristic symptoms are exophthalmos, pulsating tumor at the inner angle of the orbit, bruit, and pulsation. The most prominent symptom is the protrusion of the eyeball, which in most cases reaches a high degree. The lids become swollen, œdematous, and discolored; the conjunctiva is chemotic, and its veins are distended and tortuous. If paralysis of the ocular muscles be present, the axis of the eye deviates, and motility is interfered with. Following the distention of the superior ophthalmic vein, its circulation is reversed, and with thickened

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walls the vein becomes practically an artery, and it forms a pulsating tumor at the upper and inner angle of the orbit. This tumor is soft and reducible, is made up of a mass of pulsating and tortuous veins, and possesses a distinct bruit and thrill. Pulsation of the globe is marked, and in most cases is visible, but it can always be appreciated by palpation. The bruit is of a continuous character, and increased at each systole, and is best heard over the globe, the pulsating tumor, and the supra-orbital region. In the eye, the retinal veins are seen to be greatly distended and pulsating; hemorrhages may occur, and in some cases papilloretinitis is present. Sight may be unimpaired, but later it grows weaker, and blindness may follow. Subjectively, the symptoms are well marked, the patient complaining of severe headache, disturbing sounds in the head, and ringing in the ears, and it is generally for their relief that he seeks help. Momentary digital compression of the carotid on the affected side is followed by the disappearance of pulsation and bruit, protrusion of the bulb lessens, the pulsating tumor sinks, and the subjective symptoms are relieved, but, on removing the finger, the previous condition speedily returns. The sequence of the symptoms may vary, but, as a rule, exophthalmos appears before pulsation. The natural course of the disease, and the ultimate result of the process, if left to itself, or if not affected by treatment, is a steady increase of the symptoms, when, having reached their height, the exophthalmos, the chemosis, and the swollen lids gradually subside, but the subjective symptoms, as well as the pulsating tumor remain, and the eyesight, gradually growing weaker, is finally destroyed. In a few instances spontaneous cure has resulted. The treatment of arteriovenous aneurism of the internal carotid and cavernous sinus presents a different proposition in comparison with the treatment of a similar process in other parts of the body. The indication to be met is different, and we are limited in our choice of a method. In the extremities, for instance, in attempting a radical cure, we can shut off the flow of arterial blood into the vein by means of the double ligature, or we can make use of the quadruple ligature and extirpate the

sac. Such methods are obviously impossible in the treatment of the variety under consideration, and the problem to be solved is the reduction of the pressure in the sinus, so that the passive congestion in the superior ophthalmic vein and its branches may be relieved. The results of autopsies have added nothing conclusive concerning the healing process, but it has been represented as follows. The rent in the arterial wall is plugged with a firm clot, and, in order that it may withstand the pressure of the carotid, a reduction, or temporary abolition of the carotid pressure, is necessary. This is accomplished by ligation of the common carotid, and the blood then coagulates in the artery, as well as in the cavernous sinus, and the superior ophthalmic vein. The clot, thus resting quietly and supported on either side, gains a chance to organize, and by the time the collateral circulation is established, the healing process has reached a point of safety. Healing of the rent in the artery, in this manner, may be possible when the rent is very small and due to disease of the arterial wall, as the local conditions in the idiopathic cases are more favorable for coagulation of the blood. In the traumatic cases, however, such a healing process is not likely, as the arterial wall is healthy, its inner surface smooth and of normal consistence, accordingly the chances for coagulation are not favorable. Healing in these cases is probably from thrombosis, beginning at the superior ophthalmic vein and extending into the sinus, thus giving the necessary lateral support. When the communication between the artery and sinus is large, and rupture follows traumatism, complete closure of the opening by an organized clot is hardly to be expected, as, owing to the collateral circulation, the artery soon refills, and the pressure is greater than the clot can withstand. In many of the successful cases following ligation of the common carotid, the bruit, which disappeared on the tightening of the ligature, returned later on, and in other cases it never completely disappeared. As the bruit, on its return, was diminished in intensity, or had changed the character of its tone, it is evident that the rupture was not completely closed, but rather that the size of the opening was smaller. In these cases it is

probable that the clots deposited on the edges of the rent maintained their position through the reduction of arterial pressure by ligation, and that, by the time the collateral circulation was established, they were so far organized that the healing process could continue. When the healing was complete, the opening between the artery and sinus had narrowed down enough to allow the sinus to carry off the excess of arterial blood entering it, and thus the pressure in the sinus was not sufficient to cause passive congestion in the superior ophthalmic vein. As spontaneous cure is possible, and from the fact that a few cases have recovered under medical treatment, some authorities still advise the trial of local and internal remedies before a resort is made to surgical measures. Such advice is very unwise, since the chances of success are remote, and the time thus spent may result in the serious impairment, if not the loss, of sight. Medical treatment is indicated in cases where the patient is very old, or when the arteries are so diseased as to forbid surgical methods. In the beginning of the disease, when the symptoms are not developed sufficiently to allow of a diagnosis, rest in bed, cold applications, narcotics, etc., are indicated; but when the nature of the trouble is recognized, then surgical treatment should be promptly instituted. As to the surgical methods, we have the choice of compression or of ligation of the common carotid artery. Compression, previous to the days of antisepsis, was considered the safer method, but its results have not been satisfactory, as may be readily understood when one considers the difference which exists between a true aneurism and an arterio-venous aneurism. Its use is limited to the treatment of the idiopathic cases, where the conditions are more favorable for the deposition of clots. Compression with the finger is preferable to an instrument, but the method is always inconvenient, often painful, and its success is dependent on many conditions difficult of control. Sattler reported twenty-nine cases treated by compression, and in only four of them was there a cure. Slomann has added twenty-four cases to those reported by Sattler, making fifty-three cases in all. Of these, fifteen were cured and improved, apparently an improvement on Sattler's

figures, but in Sattler's list there were four cases improved, thus making his percentage 31 to Slomann's 27. These percentages of success are so much less than those obtained after ligation of the common carotid, that the latter method is by far the preferable one. Ligation of the common carotid artery may be said to be the operation of choice among surgeons, and as such is recommended in the surgical text-books. Pulsation and bruit, as a rule, disappear with the tying of the ligature, and the exophthalmos, the chemosis, as well as the subjective symptoms, steadily subside, and in the successful cases cure is complete from within three to six weeks. As previously stated, the bruit in many cases returns, but weaker and changed in character. The results of ligation have been satisfactory, the mortality is much less as compared with the mortality in general of the same operation, and secondary cerebral disturbances are uncommon. The mortality, in general, after ligation of the common carotid, is very considerable, probably above 25 per cent.: in exophthalmos following carotid ligation, the mortality is 10 per cent. Secondary cerebral disturbances, the greatest danger attending carotid ligation, occurred in only four of the sixty-three cases reported by Sattler, and in every instance it was an idiopathic case, and the patient a woman. The results of ligation have been collected by Sattler and by Slomann, whose work is a continuation of the former's extensive monograph. Sattler, in 1880, collected all cases hitherto reported, and they were fifty-six in number. The results were, healed, 37, or 66 per cent.; unimproved, 11, or 19 per cent.; died, 8, or 14 per cent.

Slomann, in 1898, reported ninety-five cases. Of these were, cured, 49, or 51 per cent.; improved, 17, or 17 per cent.; unimproved, 17, or 17 per cent.; died, 10, or 10 per cent.

The term "healed," as used by Sattler, includes those cured and improved. In comparing these lists, it will be seen that the percentage of cured and improved is nearly the same, but the mortality is reduced by 4 per cent. Whether the successes were permanent is uncertain, as in the majority of cases classed as "healed" the patients were lost sight of after their

discharge. Bodon, in 1899, collected all the reported cases of traumatic exophthalmos treated by ligation of the common carotid. They were fifty-eight in number, with the following results: Cured, 26, or 46 per cent.; improved, 20, or 35 per cent.; unimproved, 6, or 10 per cent.; died, 6, or 10 per cent.

The results are apparently very satisfactory, as in 80 per cent. of the cases the operation was followed by cure or improvement. In six cases blindness resulted, as the operation was done too late. The time of its performance varied from four to sixteen weeks (in one case eighteen months) after the injury. This unfortunate termination in 10 per cent. of the cases emphasizes the necessity of an early operation. Four of the six deaths were due to sepsis and hemorrhage, complications which are less liable in the future, owing to the asepsis of the present day. In comparing the results reported by Slomann with those of Bodon, it will be noticed that the proportion between those cured and those improved is much greater in Slomann's list, and its explanation is due to the fact that Bodon's list is limited to the traumatic cases. Recurrence is much more frequent in the traumatic cases: in the eleven cases of Sattler, eight were traumatic, and is due to the local condition being unfavorable for the deposition and organization of clots about the rupture in the arterial wall. The refilling of the artery, with the accompanying rise of pressure, should take place slowly and gradually, in order that the healing process should proceed satisfactorily. This process, therefore, depends on the rate at which the collateral circulation is established. The later its completion the better the chances of success. After the ligation of the common carotid artery, the collateral circulation is completely established by the free communications existing between the carotid arteries of opposite sides, both outside and inside of the skull, and through the superior thyroid artery of the side on which the carotid has been tied. The superior thyroid artery plays an important part in the establishment of the collateral circulation, and in several cases this artery was found, some weeks after the carotid ligation, to be dilated and strongly pulsating, while in the portion of the common carotid

above the ligature, and in the facial and temporal arteries, pulsation was absent. Legouest (quoted by Bodon) ligated the left common carotid in a traumatic case eight weeks after the injury. No improvement followed, and he then tied the external carotid below the origin of the superior thyroid artery. Improvement immediately followed; the bruit which was present disappeared, and cure was complete at the end of the fourth month. By cutting off the superior thyroid supply, the establishment of the collateral circulation is thereby delayed, and time is gained for the organization of the clots deposited upon the edges of the rupture. Tying the internal instead of the common carotid, therefore, offers a better prospect for success, and, in my opinion, ligation of the internal carotid artery should be the operation of choice. In a number of cases after ligation of the common carotid complete recurrence has followed, the appearance of the symptoms varying in time from a few hours up to ten weeks, in one case not until nine months had elapsed after operation. On compressing the other carotid, the pulsation and bruit disappear, and from this fact the second carotid may be ligated. Bodon, in 1899, collected all the cases on record of ligation of both common carotids for the cure of pulsating exophthalmos. He found five cases, and adding one of Dollinger's there were six cases in all. The first case was that of Gurdon Buck, in which fourteen months elapsed between the two operations. All the symptoms disappeared after the second operation, but sight of the affected eye was lost. The second case, by Foote Williams, the second operation thirty days after the first, complete cure at the end of the fourth week, sight decidedly improved. The third case, by Le Fort, fifty-four days interval, complete cure, patient perfectly well eighteen months after the last operation. The fourth was by Reeve, no mention of time interval, and the result a failure. The fifth case, by Francke, sixty days between operations, gradual improvement, cure complete at the end of a year. The sixth case, by Dollinger, first operation five months after injury, second operation thirty-two days after first, and followed by improvement. In considering these cases, one is impressed

by the fact that the operation was attended with no mortality, and, with the exception of Reeve's case, the patient was cured or improved. In Buck's case, the second operation was performed too late to save the sight, and it is a warning to us not to wait too long before performing the second operation. The ligation of both carotids is apparently not as dangerous as we would imagine, and the absence of mortality in the list reported is explained by the fact that the arteries were not diseased. The final outcome in Dollinger's case is of special interest, as it has a practical bearing on the treatment of pulsating exophthalmos. Ten weeks after the second operation, the severe headache returned, exophthalmos had increased, and at the inner angle of the orbit was a pulsating tumor, compression of which caused a complete disappearance of the bruit and a diminution of the headache. Dollinger, through an incision above the supraorbital arch, tied off several small veins, and then resected about half an inch of the enlarged and thickened superior ophthalmic vein. The bruit immediately disappeared, the headache was relieved, and thirteen days afterwards the patient was discharged cured. Woodward reports a similar experience. After ligation of the common carotid the exophthalmos and subjective symptoms disappeared, the bruit, however, returning from time to time. Ten weeks afterwards the patient returned, complaining of ringing noises in the head, which he could stop by pressure on the nose at the inner angle of the orbit. Woodward then ligated the enlarged veins at the inner and upper angle of orbit; the ringing noises disappeared, and when seen, eighteen months later, the patient was perfectly well. Cure in these two cases was probably due to thrombosis, beginning at the site of the ligature, and spreading into the cavernous sinus. These results suggest the possibility of a cure by merely tying off the mass of enlarged and pulsating veins at the inner angle of the orbit, and, if not successful, then the carotid ligation should follow. When recurrence follows ligation of the carotid, it would be well, and certainly safer, to ligate and resect the veins before attempting the ligation of the second carotid.

Slomann, in 1898, collected all the cases of pulsating exophthalmos reported up to that time, including Sattler's cases, and they were 197 in all. After excluding twenty-two cases, in which the characteristic symptoms were in the course of an non-aneurismal disease, and which Slomann designated as "false pulsating exophthalmos," there remained 175 cases of the true variety. From these figures it would appear as if the disease is not uncommon, but, as the number comprises all cases reported during the past eighty-eight years, it will be seen that the disease is rather rare. Consequently, it may be of interest to report a case which was referred to me for operation by Dr. Charles S. Bull, to whom I am indebted for the patient's history previous to his admission to my service at the New York Hospital. It is as follows:

R. M., male, twenty-nine years old, laborer, on the night of January 8, 1902, was struck on the left parietal region with a bottle. The bottle was full of beer, and was not broken by the blow. The patient was not knocked down by the blow nor rendered unconscious, and there was no laceration of the scalp. The next morning he became conscious of a continuous roaring noise in the left side of his head, which rapidly grew and extended all over his head. On the second day the left eye began to protrude and the eyelids to swell. Within a week both eyes showed enormously distended conjunctival and subconjunctival veins, and engorgement of the subcutaneous veins of both sides of both eyes, the engorgement and distention of all the vessels being more marked on the left side. Headache was severe during the first week; this soon abated, and the patient was left with a sense of confusion, which he attributed to the constant noise in his head. He applied at the New York Eye Infirmary early in February, nearly a month after the injury, and the following condition was noted. The left eye projected directly forward, and at least one-half of its anteroposterior diameter projected beyond the plane of the orbit. There was marked chemosis of the conjunctiva and great swelling of the lids, the œdema extending up under the eyebrow, downward upon the cheek, and outward towards the temple. The skin of the lids was of a dusky, purplish hue from obstruction to the venous circulation. The chemosis was most

marked downward and outward. The right eye also protruded slightly forward, but there was no chemosis. The conjunctival and subconjunctival veins of both eyes were greatly engorged and very tortuous. There was a loud bruit heard over the left eye and left side of the forehead and left temple, and this bruit was distinctly audible on the right side of the head, vertex, and occipital regions. There was a distinct pulsation of the left eyeball, perceptible to the eye as well as to the finger, but none of the right eye. There was no interference with the motility in any direction of either eye. The tension of the eyes was normal, normal pupils, and the irides of normal reaction. Vision was 20/20 in each eye, and there was no defect or limitation of the field of vision. The media were clear, and there was distinct pulsation of the retinal veins, which were greatly engorged, but without any accompanying hæmorrhages. He was admitted to the New York Hospital on February 26 for ligation of the common carotid. The symptoms had steadily increased. There was now distinct pulsation of the right eye, and above, and outward to the left eye, a mass of engorged and pulsating veins could be plainly felt. The axis of the left eye deviated outward, and its motility was impaired. On March 1 the left common carotid was ligated above the omohyoid muscle by two medium-sized chromic gut ligatures, three-fourths inch apart, the artery severed between the ligatures. Pulsation and bruit immediately ceased when the ligatures were tied. The exophthalmos, the chemosis, and swelling of the lids gradually subsided, and the patient left the hospital on the tenth day with wound healed per primam. During the stay in the hospital, the engorgement of the subconjunctival veins diminished but little, and was well marked at time of discharge. He was not seen again until February, 1903, nearly a year after operation, and there remained a slight degree of exophthalmos of the left eye, the subconjunctival veins less engorged. Patient felt perfectly well, and there was no bruit or pulsation anywhere to be discerned. The eyesight was good, the retinal veins were engorged and tortuous, and all over the retina of the left eye were the remains of numerous hæmorrhages.

In June, 1903, the patient appeared, complaining that the appearance of his eyes prevented him from securing employment, as their congested appearance suggested a free use of alcohol. Exophthalmos of the left eyeball was more marked, and the en-

gorgement of the subconjunctival veins had increased. Pulsation had returned in the external carotid and in the superior thyroid artery. A large tortuous vein of the forehead was seen; it started at about the supra-orbital notch and ran upward and outward towards the vertex. No pulsation or bruit. Beneath the vein could be felt a distinct fissure, which apparently ran into the roof of the orbit. No subjective symptoms. Further operation was advised, but the patient disappeared, and has not since been seen.

This was a case of double pulsating exophthalmos following traumatism, and apparently cured by ligation of the common carotid artery, but more than a year after operation symptoms of recurrence began to develop. As a rule, recurrence of symptoms after operation appear within a short time, generally a few weeks, and its delay for more than a year in this case is most exceptional. The protrusion of the right eyeball was undoubtedly due to rupture of each internal carotid into the cavernous sinus, and its extension by way of the circular sinus into its fellow of the right side. Double pulsating exophthalmos may be due to rupture of each internal carotid into the cavernous sinus, but generally the rupture is unilateral, and, owing to the pressure extending to the sinus of the opposite side, protrusions and pulsations of the opposite eyeball follow, but less marked than on the eyeball on the injured side. It will be noticed in this case that subconjunctival veins remained distended, while all other symptoms disappeared; and it is evident that the requisite reduction of pressure in the cavernous sinus was never obtained. With the complete return of collateral circulation, as seen by the presence of pulsation in the external carotid and superior thyroid arteries, the sinus pressure increased, and, as a result, the exophthalmos grew larger, the conjunctival congestion became more marked, and the supra-orbital vein made its appearance. The error as regards treatment was the ligation of the common carotid, and, had the internal carotid been tied, the chances of recurrence would have been lessened. Should another opportunity present itself in the future, I would tie the internal carotid. Should the patient re-

turn for operation, I would follow Woodward's example and ligate, and resect the branches of the superior ophthalmic vein at the inner angle of the orbit, and if necessary resect a portion of the main vein. In case of failure, then ligation of the opposite carotid would be indicated. Figs. 1 and 2 were taken before operation. The dotted lines in Fig. 1 indicate the area over which the bruit was audible. The crosses show the points of maximum intensity. In Fig. 2 is seen the extreme chemosis. Fig. 3 was taken a week after operation.

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FIG. 1.—Dotted line indicates area of laceration. Crosses are points of its maximum intensity.



FIG. 2.—Shows the hemostasis of conjunctiva.



FIG. 3.—One week after ligation of left common carotid.